Objectives: The aim of this EULAR Task Force was to identify a core set of data items which can easily be collected from clinicians and facilitates examination of disease course and outcome.

Methods: A multidisciplinary EULAR task force group of 20 experts including rheumatologists, epidemiologists and patient representatives was assembled and breakout groups formed for a meeting at which items from a previously compiled collection of core parameters for a GCA registry were evaluated. Results were presented to the other group members following a structured process for discussion and consensus finding. The meeting was followed by several rounds of discussions to achieve consensus.

Results: A total of 95 items were identified, subdivided into the following categories: General, Demographics, GCA-related signs and symptoms, Other medical conditions, and Treatment. Suitable instruments and assessment intervals were determined for documentation of each item. To facilitate implementation of the recommendations in both primary care and scientifically oriented registers, a minimum core set of parameters was distilled, with supplemental items that can be added optionally depending on the designated purpose of individual registers.

Abstract SAT0535 – Table 1. A minimum core set of parameters to be collected in giant cell arteritis registries and databases

Item	Instrument
General ^{1,2}	
visit date	date
Demographics	
age ³	date of birth
GCA-related Signs & Symptoms ^{3,7}	
cranial ¹	
ocular involvement	
permanent partial visual loss / blindness	y/n (examination)
transient visual loss (amaurosis fugax)	y/n (interview)
diplopia	y/n (interview)
headache	y/n (interview)
Treatment ²	
glucocorticoids ^{3,7}	
current GC use	mg per day in prednisone equivalent, route of administration
tapering ongoing	y/n
recent GC use	
continuous (> 3 months) intake	y/n (interview)
immunosuppressants	
conventional synthetic DMARDs	current medication ^{3,7}
	historical treatment ^{3,8}
biological DMARDs	current medication ^{3,7}

¹ record at baseline
² record during follow-up
³ report with date in case it occurs
⁴ record every 3-6 months; acute changes need to be recorded whenever they occur (e.g. new medication, imaging finding, osteoporotic fracture)
⁵ record every 6-12 months; acute changes need to be recorded whenever they occur (e.g. new medication, imaging finding, osteoporotic fracture)
⁶ record annually; acute changes need to be recorded whenever they occur (e.g. new medication, imaging finding, osteoporotic fracture)

Conclusions: This core set intends to ensure that data from different GCA registries and databases can be compared for the dual purposes of clinical research and improving clinical care, thereby facilitating collaborative analyses.

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SAT0536 MULTIPLE RELAPSES IN PATIENTS WITH GIANT CELL ARTERITIS

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Background: Glucocorticoids (GC) remain the mainstay of treatment for patients with GCA. However, relapses and recurrences (events) occurred in 34% to 74.5%

of the patients.¹⁻³ Most studies compared mainly patients with disease relapses/recurrences to whom without any.

Objectives: To compare patients who had multiple events to those who presented only one in order to identify characteristics and predictive factors of multiple events in patient with GCA

Methods: From 1976 to 2016, we collected prospectively clinical, laboratory, and pathological data, and the treatments and outcomes, of consecutive patients who were diagnosed to have GCA based ACR classification criteria. We compared these data in patients who had more than one event (group 1), either a relapse or a recurrence, to those who had only one (group 2). Relapse was defined as occurrence of clinical symptoms and/or inflammatory parameters, attributable to GCA, which required increased medication during follow-up i.e. under GC therapy while new disease activity after a period of remission was defined as a recurrence.

Results: We included 494 patients for whom data was collected. Forty-seven patients were excluded from our cohort due to lack of data. Among the 447 patients, 228 (51%) presented at least one event. Mean age of this population was 73.3±7 years with female predominance (70%). The median follow-up was 75±55 months. Among these patients 139 (61%) had only one event while 89 (39%) presented more than one. The total number of events was 477 of which 89 recurrences. Events occurred during the first year of treatment in 74% of patients. The majority of these events occurred with a mean prednisone dose of 12.8 ±9 mg/day (median dose 10 mg/d). In univariate analysis, significant differences between both groups is illustrated in table 1. Multivariate analysis showed that ear pain and an elevated ESR were predictive factors of multiple events (OR: 2.45 and 1.93; p=0.04 and p=0.03 respectively). We also found that this risk is lower in patients over 80 years-old (OR: 0.35, p=0.008)

Abstract SAT0536 – Table 1

Characteristic	Group 1 (>1; n=89)	Group 2 (=1; n=139)	P
Age (years)	72.67	75.34	0.011
Cough (%)	20.7	10.9	0.042
Ear pain (%)	19.5	9.4	0.029
PMR (%)	11.3	4.3	0.041
Duration of CS (m)	53.7	26.3	0.0001
Complications of CS(%)	28.4	16	0.024
Steroid sparing treatment (%)	30.7	17.8	0.024
Remission(%)	35.6	62.1	0.0001
Death(%)	23.8	36.4	0.046

Conclusions: Patients with more than 1 event represent 7% to 46% of patients with event¹⁻³ which is consistent with our study (39%). The event occurred most often during the first year of GC therapy in our cohort (74% of patients) while it affects 24% to 50% of patients in other series for the same period.¹⁻³ This difference could be explained by the heterogeneity of GC protocols. We did not found any positive correlation with hypertension, diabetes, and deep vein thrombosis that seemed to be more frequent in patients with multiple events.² In our study, this group of patients appeared younger and presented more often with cough, ear pain, and polymyalgia rheumatica that preceded GCA. Logically, in these patients, corticosteroid therapy was longer and the use of GC-sparing agent was more common. Although getting remission was more difficult in these patients, the long-term prognosis is not poor.

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SAT0537 IS BALANCE AFFECTED IN BEHCET'S DISEASE AND WHAT ARE THE FACTORS THAT EFFECT THE BALANCE?

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Background: Behcet's Disease (BD) is a multisystem vasculitis that has a broad range of manifestations. Balance as a complex task may be affected in BD and this may cause postural instability and fall risk.

Objectives: The aim of this study was to determine the fall risk in cases with BD with an objective computerised technique and to evaluate the potential related risk factors for falls in these cases.

Methods: After calculating sample size as 24 (with%95 confidence interval and± %5 standard deviation), 30 patients with BD (according to The Behcet's

Syndrome International Study Group Criteria) and 30 healthy controls were included. Cases who were not able to tolerate posturography, with a history of orthopaedic surgery and with known balance problems were excluded. The age, gender, disease duration, anamnesis of falls (last 12 months), fear of falling (yes/no) and drugs used were recorded. Also disease activity (with Behçet's Disease Current Activity Form; BDCAF) and fall efficacy (with Tinetti's Falls Efficacy Scale) were evaluated. Fall risk analysis was performed by Tetrax Interactive Balance System which is a computerised posturography device. By this method, fall risk is obtained as a numeric value (0–100) and as ranges indicating low, moderate or high risk of fall. We investigated age, gender, disease duration, fall anamnesis, fear of falling, drug usage, fall efficacy, disease activity as possible related factors to fall risk. Mann-Whitney U, chi square and Spearman correlation tests were used for statistical analysis.

Results: The mean ages of the cases and controls were 35,17±9,48 and 33,03±11,813 years, respectively. Symptom duration of the cases was 7,70±6,613 years. 7 cases (23,3%) had anamnesis of falls during the last 12 months, whereas only 8 control (26,7%) had this anamnesis ($p>0.05$). Fear of falling was reported by 43,3% of the cases and 40% of the controls ($p>0.05$). There was no significant difference between case and control groups in terms of FES-I scores (15,97±9,257 and 12,53±30,48 respectively; $p=0.599$). With the computerised system used, significantly higher fall risk results were recorded in patients with BD than the controls (50,40±24 710 and 23,13±11,811, respectively; $p<0.001$). Low, moderate and high fall risks were recorded as 30%, 33,3% and 36,7% of the cases and 70%, 30% and 0% of the controls and this distribution was also significantly worse in cases than controls ($p<0.05$). No significant correlation was found between fall risk and other factors including age, gender, disease duration, fall anamnesis, fear of falling, drug usage, disease activity except arthralgia which was significantly correlated with fall risk assessment in cases ($p<0.05$).

Conclusions: With an objective computerised technique, fall risk was found to be higher in cases with BD than controls in our study. The higher fall risk in these patients seems to be affected by joint involvement. An increased awareness of the potential fall risk and future studies investigating the possible coexisting balance problems in BD may contribute to the management.

Disclosure of Interest: None declared

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SAT0538

AORTIC INVOLVEMENT IN RELAPSING POLYCHONDRIITIS: A SYSTEMATIC REVIEW

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Background: Aortic involvement (AI) is an important complication of relapsing polychondritis (RP). Current literature is based on case reports and small case series.

Objectives: To delineate the clinical characteristics and outcome of AI in RP through a systematic literature review (SLR).

Methods: The SLR covered all English articles retrieved with relevant keyword combinations listed in PubMed until October 2017. Initially the titles and abstracts were screened by two investigators and articles considered to be relevant (involvement of the aorta and the aortic valve) were identified. Data extraction was done by the same investigators.

Results: The SLR revealed 352 papers of which 162 were discarded at the first step and a further 114 after full reading. After excluding 5 articles reporting on the same patient, we finally had 71 papers reporting 97 patients. The sex distribution was identifiable in 79 patients of whom 39 were men and 40 were women. The median age at the first symptom of RP was 31.5 years [IQR:24–43 years], the median age at RP diagnosis was 36 years [IQR: 28–43 years]; median age at the diagnosis of AI was 37 years [IQR:29–49 years]. Median duration from first RP diagnosis to AI diagnosis was less than 1 year [Range:0–21]. AI was the presenting symptom in 3 patients.

Seventy patients (73%) had involvement of thoracic aorta and 16 (23%) had involvement of the abdominal aorta. Other involved arteries were: coronary (8 patients), subclavian (5 patients), renal (3 patients), iliac (3 patients) and others (8 patients). Two patients had neurologic and renal involvement, respectively.

The most common symptoms were dyspnea (40%), followed by chest pain (13%), abdominal pain (13%) and fever (9%).

The diagnosis of AI was made during surgery in 6 patients and with different radiologic methods ranging from chest x-ray to PET-CT.

All patients excluding 1 received corticosteroids either alone or in combination with classical immunosuppressives (cyclophosphamide, azathioprine, methotrexate, mycophenolate-mofetil) or biologics (infliximab, tocilizumab, adalimumab).

The majority of the patients (53/87 patients (61%) underwent surgery including aortic graft replacement, coronary by-pass, aortic valve or mitral valve replacement operations.

16 patients died after a median follow up of 24 [IQR:11–43] months. The reasons for deaths were mainly vascular (acute myocardial infarction, heart failure,

cardiovascular operation, abdominal aorta dissection, aortic rupture and acute aortic valvular dysfunction).

Information on follow-up was known in 30 patients for a median of 18.5 months [IQR 10–48 months].

Conclusions: AI is less frequently recognised but prognostically important complication of RP. Thoracic aorta is the most frequently involved site. Surgery is needed in the majority of patients. Medical treatment is empirical and is based on glucocorticoids and immunosuppressives including biologic agents. Despite treatment, mortality is high (18% during a median of 24 months).

Disclosure of Interest: None declared

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SAT0539

THE FREQUENCY OF RECURRENT ORAL ULCERS IN FAMILY MEMBERS OF PATIENTS WITH BEHÇET'S DISEASE

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Background: It is unknown whether the observed geographic disparities in Behçet's Disease (BD) occurrence reflect primary genetic susceptibility or environmental influences within specific populations. Family aggregation studies may help to discriminate between environmental and genetic components but there are no large family surveys.

Objectives: The aim of this study was to contribute to a better understanding of the genetic aspect of familial aggregation studies in Turkey.

Methods: The study group consisted of siblings and children of 133 unrelated consecutive patients followed up at the BD outpatient clinic in FSM Hospital, Istanbul. Surveyed individuals were interviewed via telephone regarding if they had ever suffered from recurrent oral ulcerations (ROU). Children less than ten years old were surveyed via discussion with their parents. Symptoms and signs of Behçet's disease in family members were also interviewed via telephone. Subjects experience ROU, at least three episodes in one year, were invited to attend further examination, and also were asked whether anyone in their family had BD.

Results: Total 133 patients with BD (86 F, 47 M, respective mean ages: 39.9±11.7, 43.9±10.4 years) had 271 children (137 F, 134 M) and 642 siblings (319 sisters, 323 brothers, respective mean ages: 42.1±13.6, 40.7±14.4 years). All 642 siblings were contacted by telephone; 62 siblings (33 F, 29 M, respective mean ages: 17.9±9.8, 18.2±9.9 years) and 84 children (63 F, 21 M) had positive ROU history. All probands were invited, only 36% family members attended. Apart from patients with BD, 146 family members had ROU (14%). The estimated ROU rate among siblings and children was 9.6%, and 31% respectively. Among members there were 13 patients with BD (2 fathers, 1 mother, 2 children, 3 brothers, 5 sisters). Eleven of these were diagnosed at another centre. We identified 2 additional patients who met ISG criteria during the survey. Only five spouses had ROU (0.5% of members).

Conclusions: In a study from Istanbul, ROU was reported as 9.5% in the general population. In our previous study, that was conducted in an area that was reported as having the lowest prevalence rate of BD, this was estimated as 2.1% in the general population. The ROU rate in the current study for total family members was 16%, and 31% for children, which was higher than in the general population. The rate of patients with BD in the studied cohort, apart from previously diagnosed patients, were 140 per 10, 000 while the reported prevalence rates changed regionally between 20 per 10, 000 and 42 per 10, 000 in Turkey. These findings suggest that BD risk is primarily influenced by genetics rather than environmental conditions. Familial aggregation supports a genetic background for BD

Disclosure of Interest: None declared

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SAT0540

TREATMENT WITH INTRAVENOUS IMMUNOGLOBULIN IN THE VASCULITIS ANCA POSITIVE. 27 CASES STUDIED IN A SINGLE REFERENCE CENTRE

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Objectives: Intravenous immunoglobulins (IGIV) is a therapeutic alternative in vasculitis ANCA+ specially in cases of refractory or superinfection. We study the efficiency and safety in the short and long term of the IGIV in the vasculitis ANCA.

Methods: Descriptive and observational study of 27 patients with vasculitis ANCA from a reference tertiary hospital. We analysed the treatments received, the clinical and analytical variables and the indexes of activity and evolution (TABLE). Birmingham Vasculitis Activity (BVAS) was the activity index used, and for prognosis Five Factory Score (FFS). Results are indicated as mean ±SD when